Linear and whorled nevoid hypermelanosis

Sir,

Linear and whorled nevoid hypermelanosis is characterized by linear streaks of hyperpigmentation along the Blaschko's lines.[1] It is also known as 'zebra-like pigmentation'. The pigmentary anomalies become apparent in infancy or early childhood and usually remain stable, but may either increase in extent or fade over time. Histopathology suggests melanocytic hypermelanosis in most cases, but pigmentary incontinence has also been reported.

Striking homologies exist between hypomelanosis of Ito and linear and whorled nevoid hypermelanosis (LWH). In addition, several patients have been reported who display bands of both hypopigmentation and hyperpigmentation, making the distinction between hypomelanosis of Ito and LWH somewhat blurred.

Emerging opinion is that a revised nomenclature should group hypomelanosis of Ito and LWH as a single entity including hypopigmentation or hyperpigmentation along the lines of Blaschko, possible association with

extracutaneous anomalies seen in approximately 30% of patients, and somatic mosaicism in a yet undetermined but several different genes involved in human pigmentation.

A 4½ years old female child presented to the department of dermatology with multiple streaks of hyperpigmentation following the lines of Blaschko, all over the body. She was born of a full term vaginal delivery following an uneventful first pregnancy in the 27-year-old mother. The growth and development of the child was normal. Her parents and other family members were normal, and their was no history of consanguinity.

The child remained well for the first 6 weeks of life; but after 6 weeks, the mother noticed dark-colored streaks of linear and whorled pattern developing over the body of the child, involving almost the whole body; the streaks gradually became darker and darker. There was no history of erythema or vesiculobullous lesions. At about 1 year the lesions stabilized in color, and no new lesions developed after that.

Cutaneous examination revealed multiple, brown, hyperpigmented swirls and streaks along the lines of Blaschko, located symmetrically all over the body. The texture of the skin was normal over the streaks. In between the hyperpigmented streaks, the skin was normal. On the abdomen, a whorl type pattern was present; while the legs and arms had linear streaks. On the face, upper eyelids and ears were involved, but palms, soles, eyes, nails, and teeth of the child were normal. Skin biopsy showed diffuse basal cell hyperpigmentation with increase in the number of basal melanocytes. There was no incontinence of pigment, melanophages in dermis or giant melanosomes. The benign nature of the disorder was explained to the patient.

Somatic chromosomal mosaicism may present as isolated pigmentary abnormalities or multiple congenital anomalies with mental retardation.[2-4]

The differential diagnosis includes incontinentia pigmenti, epidermal nevi, and hypomelanosis of Ito. In incontinentia pigmenti,[5] the skin manifestations pass through four successive stages: vesicular, verrucous, whorl- or streak-like hyperpigmentation, and hypopigmented scars. Skin appendages, including hair and teeth, are commonly affected in incontinentia pigmenti with scarring alopecia and peg-shaped teeth. Lack of the above-mentioned features, absence of basal cell degeneration, incontinence of melanin pigment, tissue eosinophilia, and melanophages in the dermis in the skin histopathology ruled out the possibility of incontinentia pigmenti in the present case.

Epidermal nevi are often noticeable during infancy as hyperpigmented streaks along Blaschko's line, which become papillomatous and hyperkeratotic with time. Extensive skin involvement is often associated with skeletal, ocular, and nervous system anomalies. In the absence of histopathologic evidence of hyperkeratosis, acanthosis, elongation of rete ridges, papillomatosis, and occasional evidence of vacuolization, the possibility of epidermolytic hyperkeratosis was not entertained.

The scarcity of the reports prompts the present communication; and to the best of our knowledge, this is the second case reported from India.

The importance of this report lies in the fact that this condition is not only rare but also, there are very few cases of LWH reported in the literature till date.

B. K. Brar, B. B. Mahajan, Neerja Puri
Department of Dermatology and Venereology, GGS Medical College and Hospital, Faridkot-151 203, India.

Address for correspondence: Dr. B.K.Brar, C/o Brar Eye Hospital, Kotkopura, Faridkot, Punjab, India. E-mail: neerjaashu@rediffmail.com

REFERENCES

1. Loomis CA. Linear hyperpigmentation including mosaicism. Semin Cutan Med Surg 1997;16:44.
2. Davis DG, Shaw MW. An unusual human mosaic for skin pigmentation. N Engl J Med 1964;270:1384-9.
3. Griffths WA. Reticulate pigmentary disorders: A review. Clin Exp Dermatol 1984;9:439-50.
4. Verghese S, Naevlin A, Miller M, Burton BK. Mosaic trisomy 7 in a patient with pigmentary abnormalities. Am J Med Genet 1999;87:372-4.
5. Landy SJ, Donnai D. Incontinentia pigmenti (Block-Sulzberger syndrome). J Med Genet 1993;30:53-9.